



# Mid-term outcomes of cor triatriatum repair: comparison of biventricular physiology and univentricular physiology

## Original Article

Hiroyuki Nagao  and Toshikatsu Tanaka 

Department of Cardiology, Kobe Children's Hospital, Kobe, Japan

**Cite this article:** Nagao H and Tanaka T (2021) Mid-term outcomes of cor triatriatum repair: comparison of biventricular physiology and univentricular physiology. *Cardiology in the Young* **31**: 186–190. doi: [10.1017/S1047951120003595](https://doi.org/10.1017/S1047951120003595)

Received: 30 August 2020  
Revised: 24 September 2020  
Accepted: 2 October 2020  
First published online: 10 November 2020

### Keywords:

Cor triatriatum; univentricular heart; congenital heart disease

### Author for correspondence:

Hiroyuki Nagao, Department of Cardiology, Kobe Children's Hospital, 1-6-7 Minatojima-minamimachi, Chuo-ku, Kobe, Hyogo 650-0047, Japan. Tel: +81 078 945 7300.  
E-mail: [nagaohiroyuki@yahoo.com](mailto:nagaohiroyuki@yahoo.com)

### Abstract

**Background:** Cor triatriatum is the rarest of all congenital cardiac diseases, accounting for 0.1–0.4% of congenital heart diseases. Atrial septal defect is the most common associated defect; however, cor triatriatum is sometimes associated with univentricular heart. **Methods:** This single-centre retrospective study involved all patients who underwent the repair of cor triatriatum at Kobe Children's Hospital between 2000 and 2020. Twenty-four patients were required surgery. We conducted a survey of survival rate, early and late pulmonary vein stenosis in each group. **Results:** The survival rate of 5 years after cor triatriatum resection was 100% in the biventricular group and 82.1% in the univentricular group, respectively. The free rate for pulmonary stenosis of 5 years after surgery was 100% in the biventricular group and 90.0% in the univentricular group, respectively. There was no statistical difference in survival rate and 5 years free rate for pulmonary stenosis after surgery. **Conclusions:** The results showed that surgical correction offers good early and mid-term outcomes for both cor triatriatum with biventricular and univentricular physiologies.

Cor triatriatum is the rarest of all congenital cardiac diseases, accounting for 0.1–0.4% of congenital heart diseases.<sup>1</sup> It is an anomaly that has a fenestrated fibromuscular diaphragm covered by endocardium. It separates the upper common pulmonary venous chamber from the lower true left atrial cavity. The spectrum of disease correlates with the degree of obstruction caused by the membrane. Patients with significant obstruction are likely to present in infancy with symptoms resulting from pulmonary congestion and pulmonary arterial hypertension. Common presentations include failure to thrive, dyspnoea, cyanosis, or even shock.<sup>2</sup> Although cor triatriatum can be isolated cases, it is associated with other congenital cardiovascular anomalies.<sup>3</sup> Atrial septal defect is a common defect associated with cor triatriatum. Pulmonary venous anomalies and the persistent left superior caval vein are also often associated with cor triatriatum.<sup>4</sup> Cor triatriatum is often associated with biventricular heart disease, however, we sometimes recognise that cor triatriatum is associated with univentricular heart disease such as hypoplastic left heart syndrome.<sup>5–7</sup> The univentricular disease requires palliation to regulate pulmonary blood flow before or concurrently the operation of cor triatriatum. It is unknown whether the palliation procedure influences the incidence of pulmonary venous stenosis in patients with cor triatriatum. In addition, a previous study reported that the prognosis of all cor triatriatum includes biventricular and univentricular prognoses.<sup>8</sup> No study compares the prognosis associated with a univentricular physiology or biventricular physiology. Therefore, we compared the operation period, survival rate, and incidence of pulmonary venous stenosis with univentricular and biventricular in our institution.

## Materials and methods

### Ethics statement

Kobe Children's Hospital Institutional Review Board approved this retrospective study (Protocol no. 2-57), and individual written informed consent was waived.

### Patients

The 24 patients diagnosed with cor triatriatum who underwent surgical repair at our institution between March, 2000 and July, 2020 were included in this study. These patients were classified into two groups according to whether they had univentricular physiology: univentricular group (n = 13) and biventricular group (n = 11). Patients in the univentricular group, except one patient who had a primary Norwood procedure, underwent palliative surgery to adjust pulmonary blood flow, such as a Blalock–Taussig shunt or pulmonary artery banding. In our cases, there was no pulmonary venous obstruction due to the pulmonary vein itself before cor triatriatum resection. Pulmonary vein obstruction symptoms arose due to narrowing atrial communication or narrowing between the atrium and accessory chamber.

**Table 1.** Classification of cor triatriatum according to Lucas

I. Accessory atrial chamber receives all pulmonary veins and communicates with the left atrium
A. No other connections (classic cor triatriatum)
B. Other anomalous connections
1. To the right atrium directly
2. With totally anomalous pulmonary venous connection
II. Accessory atrial chamber receives all pulmonary veins and does not communicate with the left atrium
A. Anomalous connection to the right atrium directly (cardiac totally anomalous pulmonary venous connection with all pulmonary veins first draining to a venous confluence)
B. With totally anomalous pulmonary venous connection (supracardiac or infracardiac totally anomalous pulmonary venous connection)
III. Subtotal cor triatriatum
A. Accessory atrial chamber receives part of the pulmonary veins and connects to the left atrium
1. Remaining pulmonary veins connect normally
2. Remaining pulmonary veins connect anomalously (partial cor triatriatum with partially anomalous pulmonary venous connection)
B. Accessory atrial chamber receives part of the pulmonary veins and connects to the right atrium
1. Remaining pulmonary veins connect normally (partially anomalous pulmonary venous connection with anomalously connected pulmonary veins first draining to a venous confluence)
2. Remaining pulmonary veins connect anomalously (mixed totally anomalous pulmonary venous connection)

Reprint from Herlong et al.<sup>8</sup>

We used the modified classification of Lucas for the morphologic classification of cor triatriatum<sup>9</sup> (Table 1).

### Statistics

This study was a retrospective case-series study conducted at a single institution. Medical history, operative records, echocardiogram and catheterisation data, and outpatient clinical records were reviewed. Comparisons between groups were performed using the Mann–Whitney *U* test for data that were not normally distributed. A *p*-value of <0.05 was considered statistically significant. Survival analysis was performed according to the Kaplan–Meier method. Data analysis was performed using R version 3.5.1 (R: A language and environment for statistical computing; R Foundation for Statistical Computing, Vienna, Austria, <http://www.R-project.org/>).

### Results

Tables 2 and 3 show the clinical characteristics of each group. The median gestational age was 39.5 (25th–75th percentiles: 37.3–40.0) weeks, birth weight was 3.1 (2.9–3.2) kg, and the median age at the operation of cor triatriatum was 158 (113–290) days in the biventricular group. The median gestational age was 40 (38.0–40.0) weeks, birth weight was 2.9 (2.5–3.4) kg, and the median age at the operation of cor triatriatum 122 (78–822) days in the univentricular group. Gestational age, birth body weight, and age at operation of cor triatriatum were not significantly different between the groups (Tables 2 and 3).

### Morphology

Twelve patients (50%) were type IA, six patients (25%) were IB1, one patient (4%) was type IB2, four patients (17%) had type IIA, and one patient (4%) had type IIIA2 in all patients.

### Follow-up and mid-term outcomes

The median follow-up interval after cor triatriatum repair was 4.5 years (range: 0–8.8 years).

All patients experienced resection of the cor triatriatum membrane with surgery. Concurrent surgical procedures of cor triatriatum repair were performed in 18 patients (75%). In the univentricular group, four patients underwent the bidirectional Glenn procedure, three patients underwent with the Norwood procedure, two patients underwent with total cavopulmonary bypass, one patient underwent with the arch repair and main pulmonary artery banding, one patient underwent with pulmonary artery banding, one patient underwent ductus arteriosus ligation, and pulmonary artery banding. In the biventricular group, three patients had atrial septal defect closure, one had atrioventricular septal defect repair, one patient had PDA ligation, and one patient had partial anomalous pulmonary venous return repair. The survival rate of 5 years after cor triatriatum resection was 100% in the biventricular group and 82.1% in the univentricular group (Fig 1). There was no statistical difference. There was one patient who died 30 days after cor triatriatum repair and the Norwood procedure. His diagnose was hypoplastic left heart syndrome who required mechanical circulatory support for a cardiac failure during the peri-operative period and could not subsequently be weaned off. There was one late death, 1.8 years after cor triatriatum resection. His diagnose was double-outlet right ventricle, mitral valve atresia, coarctation with a univentricular circulation, and died from cardiac failure after the Norwood procedure.

No patient underwent reintervention for recurrent cor triatriatum. The free rate for pulmonary stenosis of 5 years after surgery was 100% in the biventricular group and 90.0% in the univentricular group (Fig 2). There was no statistical difference. There was one pulmonary venous stenosis, 4 months after cor triatriatum resection, and the Norwood procedure in a patient with hypoplastic left heart syndrome. When we performed cardiac catheterisation for bidirectional Glenn procedure, the left pulmonary artery

**Table 2.** Clinical characteristics of univentricular patients

No	Diagnosis	Gender	Gestational age (week)	Body weight at birth (kg)	Time of cor triatriatum repair (days)	Concurrent procedure with cor triatriatum repair
1	HLHS, mVSD, PDA	F	36	2.1	58	Norwood procedure
2	DORV, VSD, PASling, hypo arch, PDA	M	39	2.8	200	BDG
3	MS, DORV, ARSCA, hypoLV	M	36	NA	84	BDG
4	HLHS (MA, AA), restrictive PFO	M	30	1.2	78	PAB
5	MA hypoLV, DORV PAPVR	F	40	2.5	122	PDA ligation + PAB
6	MA, DORV, CoA PLSCV, restrictive PFO	F	38	3.3	3	arch repair, PAB
7	asplenia, cTGA, PA, PLSCV, ASD	M	40	3.5	277	BDG
8	MS, hypoLV, IAA, mVSD, PAPVR	M	41	3.8	85	Norwood procedure
9	DORV, MS, PA, PLSCV, CAVC	F	40	2.3	3996	BDG
10	DILV, PA, PLSCV	M	40	3.0	1658	No
11	DORV, hypoLV, CAVC, PA	F	40	3.7	942	TCPC
12	TA	F	40	3.0	822	TCPC
13	HLHS, restrictive PFO	M	38	2.7	3	Norwood procedure

ARSCA = aberrant right subclavian artery; ASD = atrial septal defect; BDG = bidirectional Glenn anastomosis; CAVC = complete atrioventricular canal; CoA = aortic coarctation; cTGA = corrected transposition of great arteries; DILV = double-inlet left ventricle; DORV = double-outlet right ventricle; HLHS = hypoplastic left heart syndrome; IAA = interrupted aortic arch; LV = left ventricular; MA = mitral valve atresia; MS = mitral valve stenosis; mVSD = muscular ventricular septal defect; PA = pulmonary atresia; PAB = pulmonary artery banding; PAPVR = partial anomalous pulmonary venous return; PA sling = pulmonary artery sling; PDA = patent ductus arteriosus; PFO = patent foramen ovale; PLSCV = patent left superior caval vein; TA = tricuspid atresia; TCPC = total cavopulmonary bypass.

**Table 3.** Clinical characteristics of biventricular patients

No	Diagnosis	Gender	Gestational age (week)	BW at birth (kg)	Time of cor triatriatum repair (days)	Concurrent procedure with cor triatriatum repair
1	ASD hypoplasticPA, PDA	F	36	2.1	75	PDA ligation
2	AVSD, PLSCV	F	40	3.2	158	AVSD repair
3	isolated cor triatriatum	M	36	1.8	235	No
4	ASD	M	40	3.1	673	ASD closure
5	ASD, mVSD	F	39	2.7	330	ASD closure
6	isolated cor triatriatum	F	41	3.2	30	No
7	AVSD, IVC interruption, polysplenia, LVOTS, DEX	F	38	3.3	5150	No
8	isolated cor triatriatum	F	40	3.5	130	No
9	PAPVR	M	NA	3.0	123	No
10	isolated cor triatriatum	M	40	3.1	103	No
11	ASD	M	37	3.2	250	No

ASD = atrial septal defect; AVSD = atrioventricular septal defect; DEX = dextrocardia; IVC = inferior caval vein; LVOTS = left ventricular outflow tract stenosis; mVSD = muscular ventricular septal defect; PA = pulmonary artery; PAPVR = partial anomalous pulmonary venous return; PDA = patent ductus arteriosus; PLSCV = patent left superior caval vein.

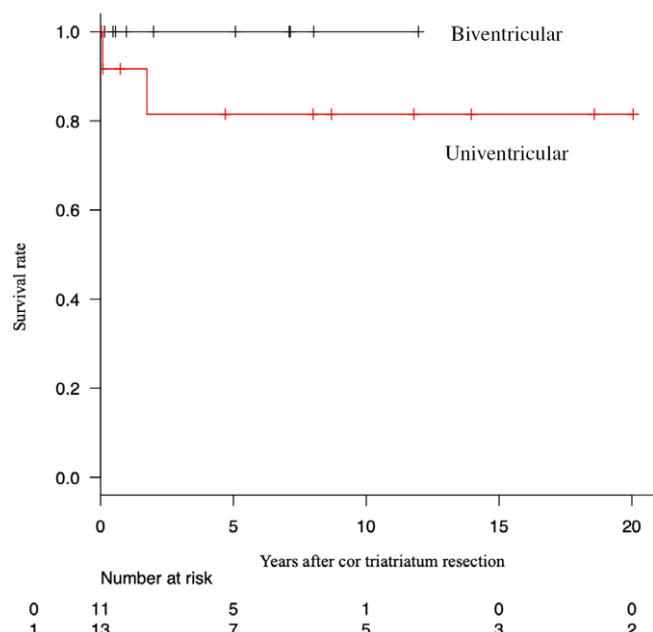
angiogram showed no left lower pulmonary vein flow. After cor triatriatum resection and the Norwood procedure, he required reintubation due to left atelectasis and phrenic nerve palsy.

In this case, although the left lower pulmonary vein was obstruction, we could not diagnose it with ultrasound before the catheter exam and collect left lower pulmonary vein catheter data. Catheter results showed that central venous pressure was 9 mmHg, right ventricular pressure was 67 mmHg, mean left upper

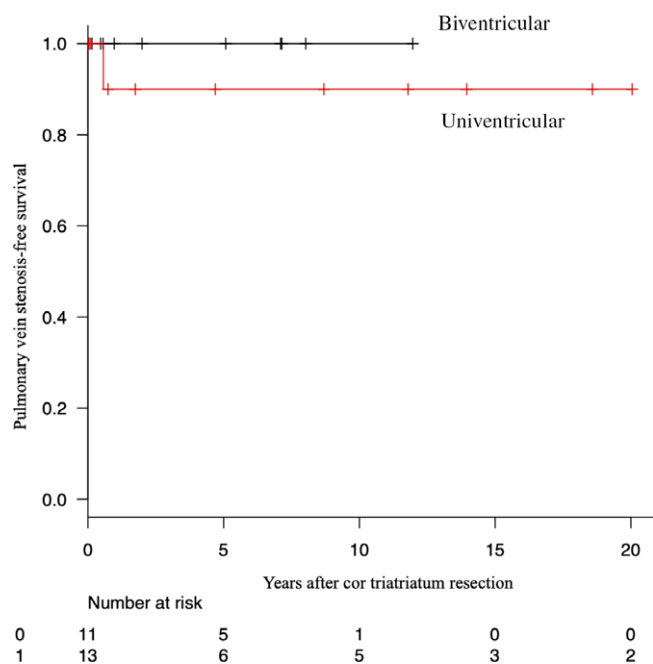
pulmonary vein pressure 11 mmHg, and mean left pulmonary artery pressure was 23 mmHg.

## Discussion

The genetics of the cor triatriatum is not completely understood. Although multiple hypotheses are proposed, the most generally accepted hypothesis, as proposed by Van Praagh et al in 1969.



**Figure 1.** Kaplan–Meier curves demonstrating survival rate over years after cor triatriatum resection.



**Figure 2.** Kaplan–Meier curves demonstrating pulmonary vein stenosis-free survival rate over years after cor triatriatum resection.

The incomplete insert of all the pulmonary veins into the left atrium causes arising in the persistence of a fibromuscular membrane between the common chamber draining pulmonary veins and the left atrium.<sup>4,10</sup>

This study has shown that cor triatriatum is associated with univentricular disease as well as biventricular disease, as seen in our institution. Although simple atrial septal defect and patent left superior caval vein are major complications associated with cor triatriatum, we should recognise that cor triatriatum may have many anatomic variants.

We diagnose cor triatriatum in various age and symptoms, and symptoms are dependent on the grade of pulmonary venous obstruction.<sup>7</sup>

Cor triatriatum remains without symptoms until adult in some cases. If the patient were diagnosed cor triatriatum incidentally with no symptoms, we should check the membrane is not restrictive at regular follow-up.<sup>6</sup>

This study showed that the median age at the operation of cor triatriatum was not different between the groups.

Palliative surgery to regulate pulmonary blood flow before the operation of cor triatriatum does not significantly influence symptoms due to cor triatriatum. Although there is no difference in survival rate, in the univentricular group, two patients died. Both patients underwent the Norwood procedure. It seems that cor triatriatum does not influence mortality directly after surgery.

However, it has a broad spectrum of presentations; infants can present with shock, pulmonary oedema, respiratory failure, and pulmonary hypertension in the most severe forms of this condition.<sup>2,11</sup>

It might raise mortality if the pre-surgery condition is an unwell such as shock even if isolated cor triatriatum. Most reported deaths occurred in children who presented in a critically ill condition, those with complex associated anomalies, or after a misdiagnosis.<sup>6</sup>

Pulmonary vein stenosis was present in 4% of patients (n = 1) after cor triatriatum repair in our study; however, there is no recurrence of the left atrial membrane in patients.

Diagnosis is usually established by echocardiography; in our case, all patients were diagnosed by echocardiography.<sup>12,13</sup>

However, cor triatriatum has various patterns; contrast CT is more useful for recognising the detailed structure of cor triatriatum and underlying disease in complex heart disease patients. The standard treatment for cor triatriatum is surgery with excision of the membrane. We performed surgery to repair in all patients in our case.

Recently, successful percutaneous balloon dilatation of cor triatriatum has also been reported. There are no guidelines that are appropriate for percutaneous or surgical treatment.<sup>14,15</sup>

It seems that percutaneous balloon dilatation is a more reasonable treatment modality for isolated cor triatriatum; thus, avoiding thoracotomy, use of extracorporeal circulation, and cardioplegic arrest of the heart in these patients. It is associated with minimal mortality and morbidity.<sup>16</sup>

However, long-term data on morbidity and mortality using the percutaneous approach are unknown.

The results showed that surgical correction offers good early and mid-term outcomes for both cor triatriatum with biventricular and univentricular physiologies. The age at operation of cor triatriatum was not statistically different between the groups.

**Financial support.** This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

**Conflicts of interest.** None.

**References**

- Richardson JV, Doty DB, Siewers RD, Zuberbuhler JR. Cor triatriatum (subdivided left atrium). *J Thorac Cardiovasc Surg* 1981; 81: 232–238.
- Burger C. A rare case of cor triatriatum leading to respiratory failure and shock in an 8-month-old boy. *Pediatr Emerg Care* 2019; 35: e6–e8.
- Yaroglu Kazanci S, Emami S, McElhinney DB. Outcome after repair of cor triatriatum. *Am J Cardiol* 2012; 109: 412–416.

4. Kumar V, Singh RS, Mishra AK, Thingnam SKS. Surgical experience with cor triatriatum repair beyond infancy. *J Card Surg* 2019; 34: 1445–1451.
5. Naito Y, Harada Y, Uchita S, et al. Successful surgical treatment of hypoplastic left heart syndrome associated with a divided left atrium that was diagnosed intraoperatively. *J Thorac Cardiovasc Surg* 2007; 133: 813–815.
6. Alphonso N, Nørgaard MA, Newcomb A, d'Udekem Y, Brizard CP, Cochrane A. Cor triatriatum: presentation, diagnosis and long-term surgical results. *Ann Thorac Surg* 2005; 80: 1666–1671.
7. Humpl T, Reineker K, Manlhiot C, Dipchand AI, Coles JG, McCrindle BW. Cor triatriatum sinistrum in childhood: a single institution's experience. *Can J Cardiol* 2010; 26: 371–376.
8. Saxena P, Burkhart HM, Schaff HV, Daly R, Joyce LD, Dearani JA. Surgical repair of cor triatriatum sinister: the Mayo Clinic 50-year experience. *Ann Thorac Surg* 2014; 97: 1659–1663.
9. Herlong JR, Jagers JJ, Ungerleider RM. Congenital heart surgery nomenclature and database project: pulmonary venous anomalies. *Ann Thorac Surg* 2000; 69: S56–S69.
10. Van Praagh R, Corsini I. Cor triatriatum: pathologic anatomy and a consideration of morphogenesis based on 13 postmortem cases and a study of normal development of the pulmonary vein and atrial septum in 83 human embryos. *Am Heart J* 1969; 78: 379–405.
11. Gheissari A, Malm JR, Bowman FO, Bierman FZ. Cor triatriatum sinistrum: one institution's 28-year experience. *Pediatr Cardiol* 1992; 13: 85–88.
12. Thakrar A, Shapiro MD, Jassal DS, Neilan TG, King ME, Abbara S. Cor triatriatum: the utility of cardiovascular imaging. *Can J Cardiol* 2007; 23: 143–145.
13. Hamdan R, Mirochnik N, Celermajer D, Nassar P, Iserin L. Cor triatriatum sinister diagnosed in adult life with three dimensional transesophageal echocardiography. *BMC Cardiovasc Disord* 2010; 10: 54.
14. Huang TC, Lee CL, Lin CC, Tseng CJ, Hsieh KS. Use of Inoue balloon dilatation method for treatment of Cor triatriatum stenosis in a child. *Catheter Cardiovasc Interv* 2002; 57: 252–256.
15. Schiller O, Burns KM, Sinha P, Cummings SD. Cor triatriatum with partial anomalous pulmonary venous return: a rare case of parallel obstruction and successful staged treatment. *Pediatr Cardiol* 2012; 33: 363–365.
16. Li WW, Koolbergen DR, Bouma BJ, Hazekamp MG, de Mol BA, de Winter RJ. Catheter-based interventional strategies for cor triatriatum in the adult – feasibility study through a hybrid approach. *BMC Cardiovasc Disord* 2015; 15: 68.